PULMONARY INFARCTION IN A SICKLE CELL TRAIT PATIENT-PRESENTING AS ACUTE CHEST SYNDROME

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ABSTRACT

BACKGROUND

Sickle cell trait is usually asymptomatic. Anaemia and painful crises are rare. An uncommon, but highly distinctive symptom is painless haematuria probably due to papillary necrosis. Few isolated cases of massive sickling or sudden death due to exposure to high altitudes or extremes of exercise and dehydration are also reported. Though pulmonary infarction is a well-known complication of sickle cell disease, it is very uncommon in sickle cell trait. Our case is an unique presentation of a 34-year-old female who is a known sickle cell trait patient who presented as pneumonia and subsequently found to have pulmonary infarction. Patient was successfully treated with antibiotics, low molecular weight heparin (LMWH) and anticoagulants (Acitrom).

KEYWORDS

Sickle Cell Trait, Pulmonary Infarction, Acute Chest Syndrome, CT Angio.

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BACKGROUND

Sickle Cell Disease (SCD) is one of the most prevalent genetic disorders. There are more than 200 million carriers of sickle cell trait worldwide. Sickle cell disease is an autosomal recessive disorder where valine replaces glutamic acid in number 6 position of beta globin chain due to mutation. Two acute complications of sickle cell disease are Vaso-Occlusive Crisis (VOC) and Acute Chest Syndrome (ACS), but it is very rare in sickle cell trait. Isolated cases of massive sickling or sudden death due to exposure to high altitudes or extremes of exercise and dehydration has been reported. The ACS is a common complication of the sickling disorders (HbSS, HbSC, HbS ß$^+$/thalassaemia, HbS/ß-thalassaemia, etc.) and is responsible for considerable morbidity and mortality in these patients[1,2] with an incidence of 10,500/1,00,000 patients/year.[2]

CASE REPORT

A 34-year-old female known sickle cell trait was admitted with chief complaint of fever-15 days, scanty productive cough and chest pain for 10 days. She was earlier treated with antibiotics outside, but without improvement. On examination, patient was febrile, tachypneic and had bilateral crepitations on auscultation in infraaxillary and infrascapular areas. On further evaluation was found to have non-homogenous opacity in left lower zone, sputum was negative for AFB and sputum and blood culture was negative.TLC-22330 with 65% neutrophils. Patient was treated with antibiotics, oxygen and folic acid. Fever subsided after 10 days, but chest pain and tachypnoea persisted for which CT chest was done, which showed dense wedge-shaped opacity over left lower zone suggestive of a consolidation or an infarct. Subsequently, CT pulmonary angiogram was done, which showed large saddle thrombus in the division of pulmonary artery causing pulmonary infarction (left). ECG showed T-inversion in V1-V3 and 2D echo was normal. Protein-C, S, antithrombin III, serum APLA, serum homocysteine and Doppler USG leg vessels was normal. In view of all these findings, a diagnosis of pulmonary infarction in a sickle cell trait patient was made. Subsequently, patient was treated with Inj. LMWH 0.6 cc b.i.d. for 5 days along with Tab. Acitrom 2 mg once a day and monitored with PT/INR regularly. Patient symptomatically improved (chest pain and dyspnoea) and was discharged with advice of regular followup.

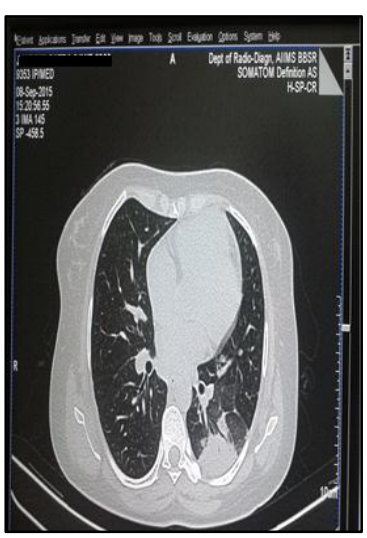

Figure 1. CT Scan Chest (8.9.15) Dense Wedge-Shaped Opacity over left lower Zone

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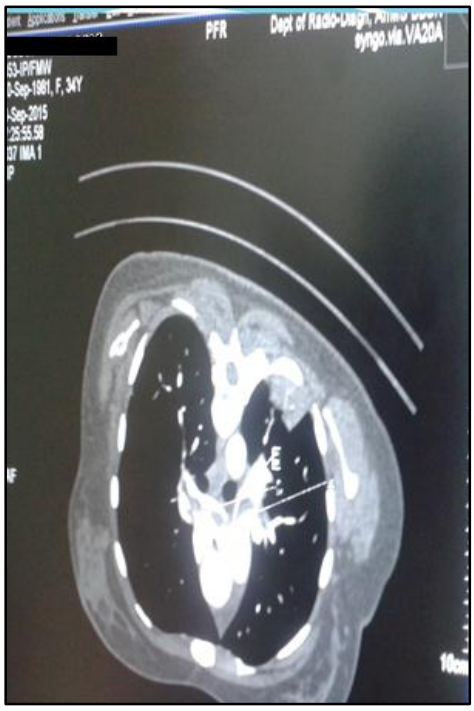

Figure 2. CT PA (10.9.15) Large Saddle Thrombus in Division of Pulmonary Artery Causing Pulmonary Infarction (Left)

DISCUSSION

The Acute Chest Syndrome (ACS) in Sickle Cell Disease (SCD) can be defined as-

1. A new infiltrate on chest x-ray.
2. Associated with one or more new symptoms- Fever, cough, sputum production, dyspnoea or hypoxia.

The symptom complex may be varied and not all symptoms are present in every episode; however, some combination of these symptoms is required for this "diagnosis." The ACS is a common complication of the sickling disorders (HbSS, HbSC, HbS/ß$^{+}$-thalassaemia, HbS/ß-thalassaemia, etc.) and is responsible for considerable morbidity and mortality in these patients.[1,2]Various aetiology includes pulmonary infarction in situ sickling, fat embolism, hypoventilation due to rib infarction and various infections. The mechanism responsible for ACS is not fully understood. It is believed to be a specific form of acute lung injury that can progress to acute respiratory distress syndrome.[3,4] The lung injury is caused by various insults superimposed on the genetically-based pathophysiology of SCD. More recent data suggest that abnormalities in endothelial cell Nitric Oxide (NO) production and metabolism as well as oxidant status may contribute to the development of ACS.[4,5,6] Clinical presentation include fever, chest pain, tachypnoea, crepitation on chest auscultations and infiltrates in chest x-ray. A high index of suspicion is necessary as this disease can be catastrophic. CT pulmonary angio is necessary for diagnosis, which typically shows thrombus in the pulmonary artery. After ruling out other hypercoagulable state, ACS diagnosis can be made. Repeated episodes of acute chest pain correlate with reduced survival. Acutely, reduction in arterial oxygen saturation is especially ominous because it promotes sickling on a massive scale. Chronic acute or subacute pulmonary crisis lead to pulmonary hypertension and cor pulmonale, an increasingly common cause of death as patients survive longer. Sickle cell trait has been characterised as a benign condition. However, life-threatening complications sometimes develop. Acute Chest Syndrome (ACS) is usually described in homozygous sickle cell disease, but it rarely develops in individuals with sickle cell trait. Treatment modalities include antibiotics, oxygen, LMWH, Acitrom, corticosteroids and hydroxyurea.

CONCLUSION

Although, acute chest syndrome is a known complication of sickle cell disease and is uncommon in sickle cell trait one has to be vigilant not to miss it because of its catastrophic nature. Any sickle cell trait, patient presented with fever, tachypnoea and chest infiltrates should do CT angio pulmonary to rule out pulmonary infarction.

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